Prenatal Diagnosis of Unusual Fetal Pial Arteriovenous Malformation

A Case Report

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Summary

Cerebral arteriovenous malformations (C-AVMs) are rarely diagnosed in utero. Most prenatal imaging of intracranial vascular malformations relates to Vein of Galen aneurysmal malformations (VGAMs) or Dural Arteriovenous Malformations (D-AVMs). We report a case of a fetal pial AVF with multiple fistulae and venous pouches, which appeared as an anechoic lesion on the prenatal ultrasound scan. The patient was asymptomatic with normal postnatal growth. No haemodynmaic disturbance was evident. Postnatal Computed tomography (CT), Magnetic Resonance Imaging (MRI) and catheter Digital Subtraction Angiography (DSA) confirmed the presence of a pial AVF. The angiographic findings and family history of nose bleeds suggests the diagnosis of Hereditary Hemorrhagic Telangiectasia. The largest AVF was embolized with tissue adhesive; the residual AVF subsequently removed by surgical excision.

Introduction

Sonographic finding of intracranial arteriovenous shunts in the fetus mostly involve the vein of Galen [1,2]. In utero diagnosis of cerebral AVMs is rare. We describe a case of a pial arteriovenous shunt of the middle cerebral arteries with a prominent venous pouch that was detected on prenatal ultrasound. The management of this asymptomatic congenital cerebral AVMs included embolization.

Case Report

A 27-year-old female on antenatal ultrasound (during the third trimester) was found to have a homogeneous round anechoic lesion along the brain's surface at the level of the right sylvian fissure (figure 1). There was no other cerebral or cardiac abnormality noted on the ultrasound scan. Since the anechoic lesion simulated a cyst, which was superficially located, the diagnosis of arachnoid cyst was made. The baby had a normal in utero development and was delivered uneventfully. Postnatal follow-up CT and MRI/MRA scan were performed when she was 15 months old. It was shown that there was a large vascular pouch along the right Sylvian fissure with feeding arteries from the right middle cerebral artery (MCA) and venous drainage to the right sphenoparietal veins. The large venous pouch corresponded exactly to the anechoic lesion on the prenatal ultrasound scan (figure 2). Associated mass effect with some degree of surrounding cerebral atrophy was also demonstrated. A pial arteriovenous fistula was then diagnosed. Catheter digital subtraction angiography (DSA) confirmed there were two venous

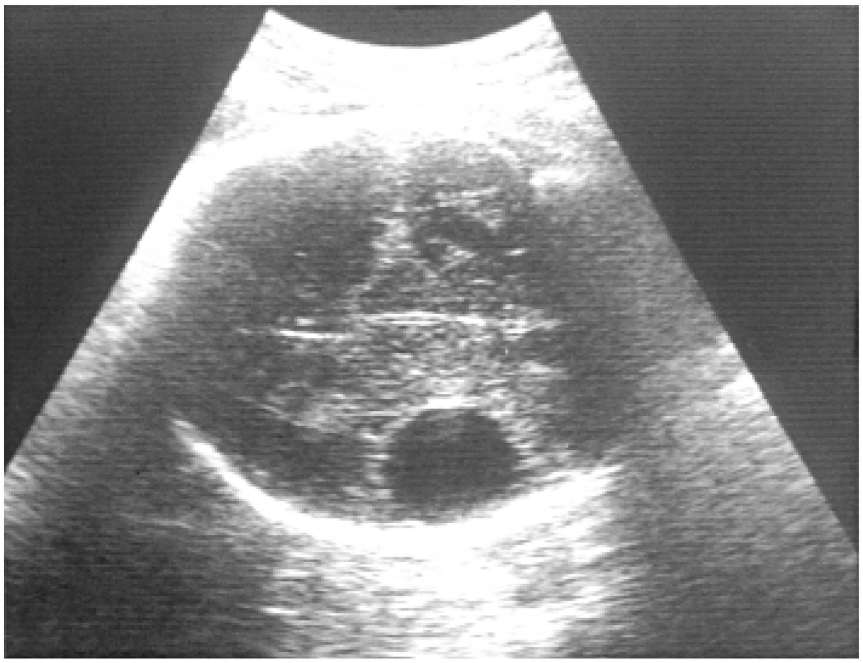

Figure 1 Prenatal ultrasound during the third trimester performed at the level of third ventricle depicts a homogeneous anechoic extraaxial lesion at the right Sylvian fissure with slight compression on the right operculum.

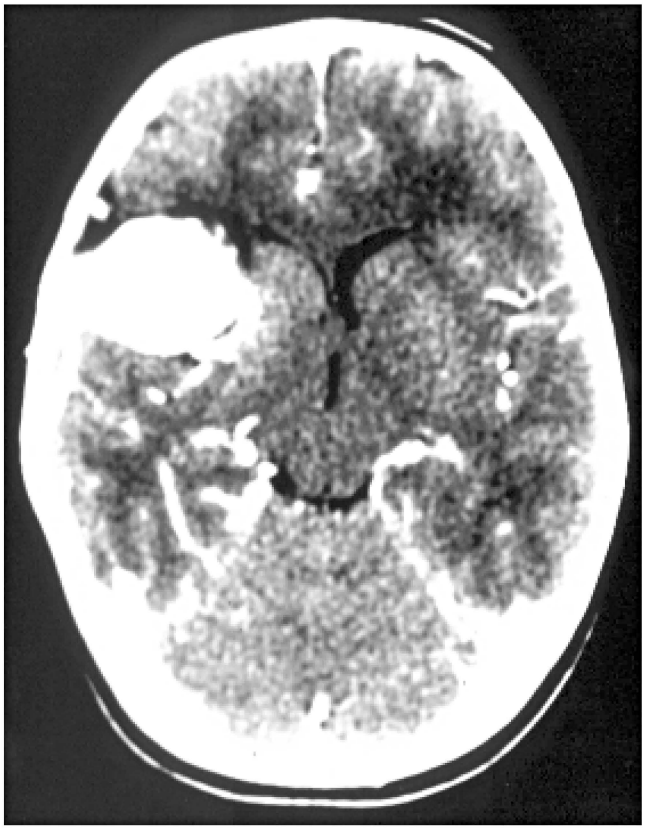

Figure 2 Postnatal post-contrast CT scan demonstrates an intense and homogeneous contrast-filling of a mass lesion located within the right Sylvian fissure similar in location and appearance to the prenatal ultrasound scan.

pouches being fed by two branches of the right MCA (figures 3, 4). The larger feeder was a single branch, which emptied directly into the larger pouch. The second feeder originated from the M2 segment of the middle cerebral artery and supplied another adjacent smaller

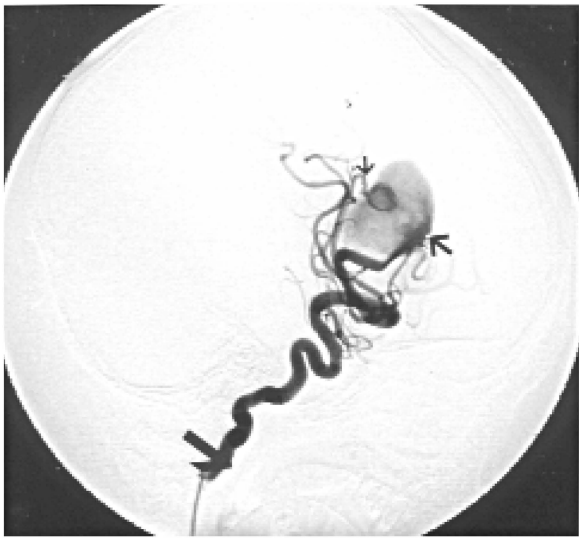

Figure 3 Pre-embolization digital subtraction angiograms (DSA) in the early arterial phase show that there are two venous pouches supplied by two separate arterial feeders from the right middle cerebral artery. The larger inferior feeder (large arrow) supplies the larger pouch while the smaller feeder (arrow) enters the smaller pouch.

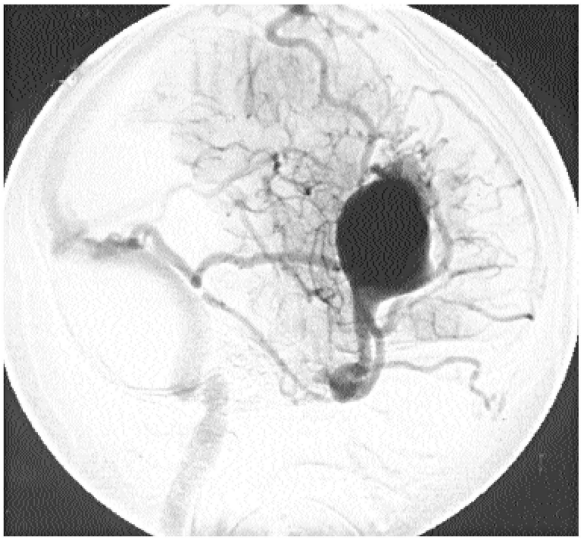

Figure 4 The venous phase of pre-embolization DSA well demonstrates the large venous pouch and the dilated draining veins through the right Sylvian vein and cortical draining veins including the vein of Trolard and vein of Labbe.

pouch. The venous drainage was cortical, involving a large descending Sylvian vein, which then ran anteromedially towards the cavernous sinus as well as laterally to the vein of Trolard and vein of Labbe. In view of the risk of future hemorrhage, embolization was performed when the child was one year old. The lower main feeder was cannulated superselectively and embolized successfully by using tissue adhesive (NBCA) (figures 5, 6, 7). The smaller upper feeder into a small venous pouch was not treated. A second embolization was attempted to obliterate the residual AVF when the patient was 3 years old but was unsuccessful. Craniotomy and excision of the residual AVF was performed when the patient was 4 years old. The patient did well post-operatively and no resi-dual AVF was demonstrated on the follow-up DSA.

Discussion

Cerebral arteriovenous malformations are considered to be congenital in nature. Most theories attribute them to either persistence of a primitive arteriovenous connection or development of such a connection after its initial closure [3]. Mullan and colleagues showed that arteriovenous malformations are generally not diagnosed in utero, suggesting that they either are too small to be detected with perinatal ultrasound in these early stages, or they actually develop after birth [4]. Our case report differs from Mullan's suggestions.

Lasjaunias [5], on the other hand, emphasized that arteriovenous malformations are the result of biological dysfunction of the remodeling process at the junction of capillaries and veins. Construction of a vascular structure with morphological and physical maturation is the result of complex biological factors and events starting in the embryo and continuing in the fetus, neonate and young infant. The vascular structures are maintained, repaired and modified in accordance with metabolic demand (via the feedback) and genetic control. The alterations in program will result in different reconstruction. Therefore, abnormal vascular structures could be related either to construction failure or failure in the renewal process, which function until the cell lines become committed to a certain cell type. If a mutation occurs during embryogenesis, arteriovenous malformations may form. Furthermore the earlier a causative event occurs the larger the area of impact, and the higher the chances of apparent multifocali-

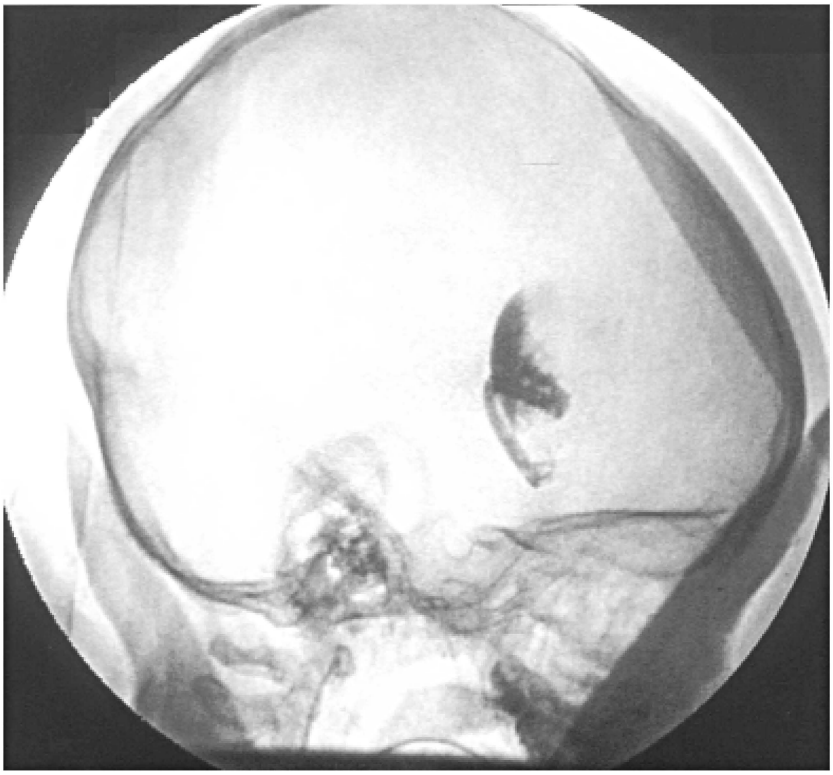

Figure 5: The post-embolization non-subtracted lateral skull view depicts the glue-cast in the venous pouch and in the feeder artery.

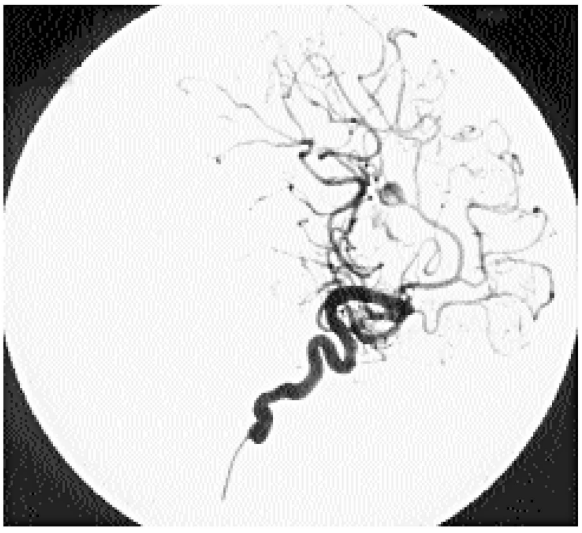

Figure 6 Post-embolization cerebral DSA in early arterial phase confirms the obliteration of the large venous pouch and its feeder and the persistent filling of the smaller venous pouch just superior to the larger pouch.

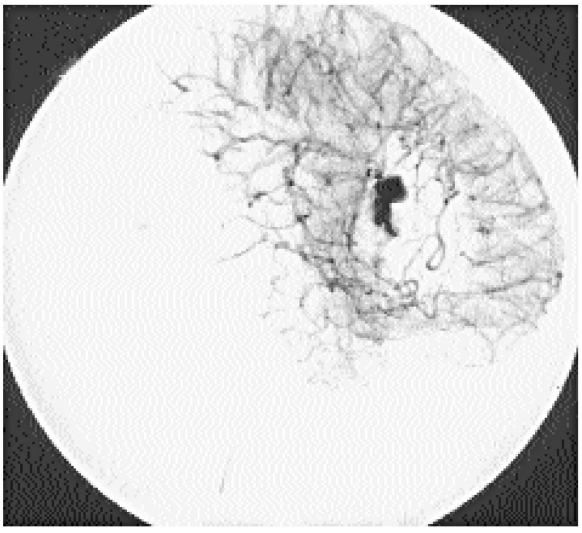

Figure 7 Post-embolization cerebral DSA in venous phase shows the filling of the smaller venous pouch. The abnormal engorged venous drainages in the pre-embolization scan had disappeared.

ty will be. The later the trigger occurs the more focal the defect and the smaller the lesion. Accordingly, growth of AVM should not occur. A large nidus will not result from the growth of a small one but rather the trigger fires on a defect shared by a larger group of cells. The "AVM growth" could be attributed to the peripheral angiogenesis or high flow angiopathic

changes. Therefore, cerebral AVMs are the result of a congenital event, which is present but occult at birth and becomes apparent later in life subsequent to "revealing triggers". The "revealing triggers" are unknown but could be mechanical, hormonal, pharmaceutical, haemodynamic, thermal, radiation, viral, infective, metabolic factors, etc. The target is likely the venous side of the capillaries (venous endothelium of the developing choroids plexus for vein of Galen aneurysmal malformation and dural venous sinus for dural sinus malformation).

Lasjaunias demonstrated the antenatal presence of only two vascular lesions: the vein of Galen aneurysmal malformation and the dural sinus malformation [5]. We propose to add B-AVM to the antenatally diagnosable vascular lesions. The very rare pial AVMs that may present in the fetal period are probably revealed by a trigger, which leads to subpial venous endothelial cell dysfunction during in utero life.

Cerebral AVMs of the fetus and neonate are rare. Even so, Comstock stated that almost all cerebral AVMs in the fetus involve the vein of Galen, although occasionally they may involve the frontal area of the brain instead [1]. There are only two reported cases of prenatal diagnosis of cerebral AVMs apart from the Vein of Galen malformation in the English literatures. Comstock reported a 5cm AVM of the left frontal lobe detected by 34 weeks [1] with hydrocephalus and generalized cardiomegaly. Paladini described the other arteriovenous fistula at 23 weeks gestation involving the middle and posterior cerebral arteries with venous aneurysm and increased cardiac output [7]. In those cases, the fetus succumbed due to termination of pregnancy or congestive heart failure after birth respectively with no other imaging performed. Our case further illustrates that not all the congenital cerebral AVFs had haemodynamic impact with poor prognosis. Our patient had an uneventful delivery and normal growth in the neonatal period. No hydrocephalus or congestive cardiac failure developed. Although an AV shunt in the brain was present it did not lead to a haemodynamic disturbance as it can do in the vein of Galen malformation. This was probably related to the fact that there were narrowed venous outlets restricting the venous outflow. Moreover, the associated draining cortical veins and venous efferents were distant from the major venous dural sinuses. The arterial pressure and flow was likely buffered before reaching the major venous outflow. The prognosis is better for those neonates who have no evidence of heart failure at birth [8]. Absence of increased hemodynamic circulation will definitely improve the prognosis.

Matsubara et al [9] stated that multiple, cortical, micro AVMs or AVFs harbouring single feeding arteries and single veins should raise a clinical suspicion of HHT-related AVMs. Furthermore, it was noted that all AVF lesions had venous pouches. Lasjaunias also reported that children with a HHT may have one or several single hole brain AVFs with large venous ectasias. Moreover, none of these patients presented with congestive cardiac failure, hydrocephalus or melting brain syndrome [5,6]. There is a strong angiographic similarity between the findings in HHT and in our case. Interestingly, both parents had a personal history of frequent nose bleeds during childhood, requiring the father being cauterized as a youngster. Matsubara et al in their discussion further pointed out that children with HHT do not always have the typical symptoms seen in adults such as mucocutaneous telangiectasia, frequent episodes of epitaxis and gastrointestinal bleeding. The first clinical presentation of HHT in a child may be a cerebral AVM. Even though the patient has no history of previous hemorrhage, other AVM or abnormal chest radiograph, Hereditary Hemorrhagic Telangiectasia (HHT) will have to be considered, especially in the presence of a probable family history.

From the diagnostic point of view, Dan suggested that colour Doppler Imaging is the most appealing method for in utero diagnosis of fetal AVMs [10]. It is a good method to differentiate cystic from vascular lesions and does make early diagnosis of cerebral AVMs possible. The in utero diagnosis could provide information on pregnancy surveillance, site of delivery, and appropriate postnatal therapy. The continuous assessment of cardiac failure or hydrocephalus, which are important prognostic indicators, could then also be addressed. Moreover, early recognition of this lesion may benefit those patients who wish to consider the option of pregnancy termination prior to 24 weeks.

Conclusion

We report a rare case of pial cortical AVFs with large venous pouches, diagnosed in utero, remaining asymptomatic after birth and treated with successful combined endovascular and surgical approaches during postnatal life. The etiology of congenital AVMs is discussed and the possibility of HHT is explored.

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